CASE REPORT

Congenital Dermoid Cyst of the Middle Ear

Paul E. Farris, M.D., William L. Meyerhoff, M.D.,
and Frank Vuitch, M.D.

Abstract—Dermoid cysts of the head and neck are rare lesions comprised of epidermal and mesodermal elements. We report a dermoid cyst presenting in the middle ear of the youngest patient reported to date. Structures of endodermal descent were also identified, but, given that the entire middle ear mucosa is of endodermal origin, specific classification as a teratoma would be imprecise. This lesion is interesting in that it did not directly involve the mastoid. Possible embryologic sites of origin are discussed.

Dermoid cysts of the head and neck are uncommon, with only 6.9% of all dermoids presenting in this area. In the classic work by New and Erich,[1] 49.5% of head and neck dermoids were located in the orbit and no lesions were specifically identified in the middle ear. Dermoids must be differentiated from teratoma, which can on occasion be malignant when located in the abdomen or retroperitoneum. Malignant teratomas of the neck have been reported in both children and adults.[2] Dermoid tumors may present as unremitting serous otitis or recurrent otitis media. Inspection will usually reveal a pale middle ear mass. Distinction from congenital cholesteatoma is impossible preoperatively but not especially important as both lesions are treated through the same approach (tympanomastoidectomy).

CASE REPORT

E. J. was an 8-month-old female infant who was referred for evaluation of a pale left middle ear mass. There was no history of otorrhea, trauma, or associated congenital anomaly. Her physical examination was negative for evidence of facial nerve paresis, auricular or preauricular masses, facial asymmetry, or ocular findings. A computed tomographic (CT) scan of the temporal bones revealed normal anatomy on the right but an abnormal soft tissue density filling the left mesotympanum and hypotympanum (Figs. 1–3). Bony expansion of the hypotympanum was noted. Eustachian tube extension and expansion was seen, as was continuity with the tympanic membrane. The ossicles were completely encased. No facial canal, labyrinthine, or tegmen erosion was seen (Fig. 3). These findings were initially deemed consistent with congenital cholesteatoma, which was the working diagnosis. Attempts were made to acquire soundfield threshold data. Localization was elicited at 20 dB on the right and 40 dB on the left. Auditory brainstem evoked response testing was consistent with a moderate conductive hearing loss on the left.

E. J. underwent canal wall-up tympanomastoidectomy, which revealed yellow fluid filling the mastoid, suggestive of cholesterol granuloma. A mass was identified in the protympanum with extension into the middle ear, nearly filling the cleft. The incus was eroded in several areas. The incudostapedial joint was dislocated by the mass, which encased the malleus also. After some effort a 2.5 × 2 cm firm, white soft tissue mass was delivered. A segment of the mass extended into and expanded the proximal 1 cm of the eustachian tube. The stapes footplate was mobile.

E. J. did well and underwent revision with ossiculoplasty eight months later. An anterior oval window perilymph leak was identified and grafted. No recurrence was seen. She continues to do well three years later.

Skull Base Surgery, Volume 8, Number 2, 1988 Department of Otolaryngology—Head and Neck Surgery, University of Arkansas for Medical Sciences, Little Rock, Arkansas (P.F.), and Departments of Otolaryngology—Head and Neck Surgery (W.M.) and Pathology (F.W.), University of Texas Southwestern Medical School, Dallas, Texas Reprint requests: Dr. Farris, Department of Otolaryngology—Head and Neck Surgery, University of Arkansas for Medical Sciences, 4301 West Markham St., Slot 543, Little Rock, AR 72205 Copyright © 1998 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001. All rights reserved.

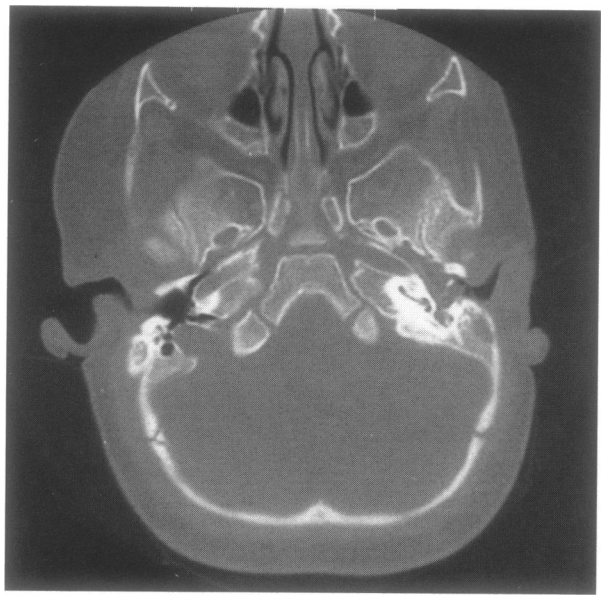

Figure 1. Axial CT of the skull base showing left middle ear opacification and expansion of the proximal one third of the eustachian tube.

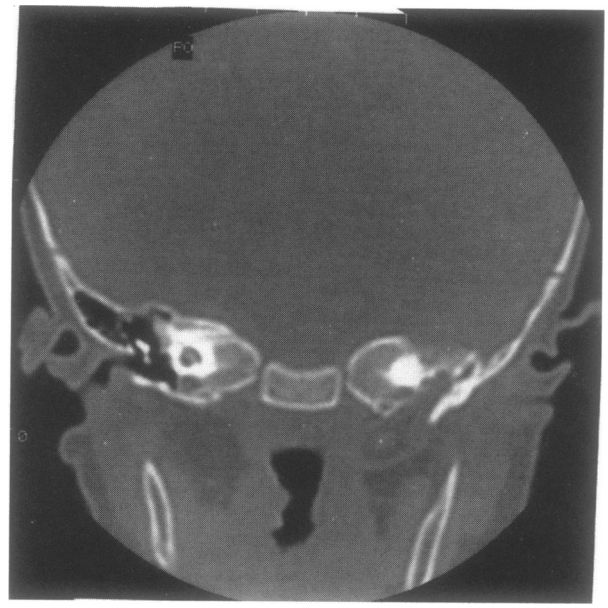

Figure 3. Coronal CT demonstrating proximal eustachian tube expansion.

PATHOLOGY

Histopathological examination revealed a benign neoplasm with areas of pseudostratified columnar epithelium, respiratory epithelium in a bronchiolar arrangement, seromucinous glandular tissue consistent with salivary gland, as well as bone, hair follicles, and metaplastic transitional epithelium. These tissues are of primarily ectodermal and mesodermal origin. The presence of bronchiolar glands as a possible endodermal component is intriguing but difficult to interpret, as the entire middle ear mucosal surface is of endodermal origin (Figs. 4, 5). The bronchiolar gland arrangement may be a secondary change in otherwise normal middle ear mucosa. Strict criteria for classification as a teratoma are therefore not met. The presence of salivary tissue in the middle ear is probably attributable to a coexistent choristoma.

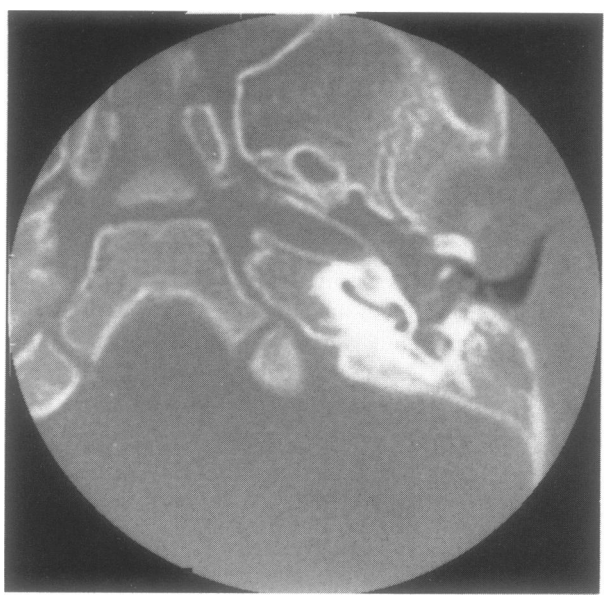

Figure 2. Magnified axial CT showing the proximity of the mass to the horizontal carotid canal, cochlear promontory, and foramen spinosum.

DISCUSSION

The first case of isolated middle ear dermoid was presented in 1866 by Hinton.[3] Five additional dermoid cysts of the middle ear and mastoid have been described since 1866 and are well outlined by Fried and Vernick.[4] Prior to their report, the most recent dermoid of the middle ear and mastoid had been reported by Steel.[5] Howie[6] described an interesting lesion presenting as a red, pea-sized mass in the middle ear of a 29-year-old woman. The presence of this mass in the hypotympanum suggested a glomus tumor. Ultimately, a dermoid associated with a dehiscent jugular bulb was identified.

Embryological sites of origin in the head and neck include the derivatives of the anterior neuropore and the region of the first branchial groove and cleft. A process of congenital inclusion has been proposed as the mechanism by which pluripotential embryonic cells are enclaved at embryologic fusion points to develop as teratomas. This process holds especially well for dermoids of the head and neck, as these can often be found at su-

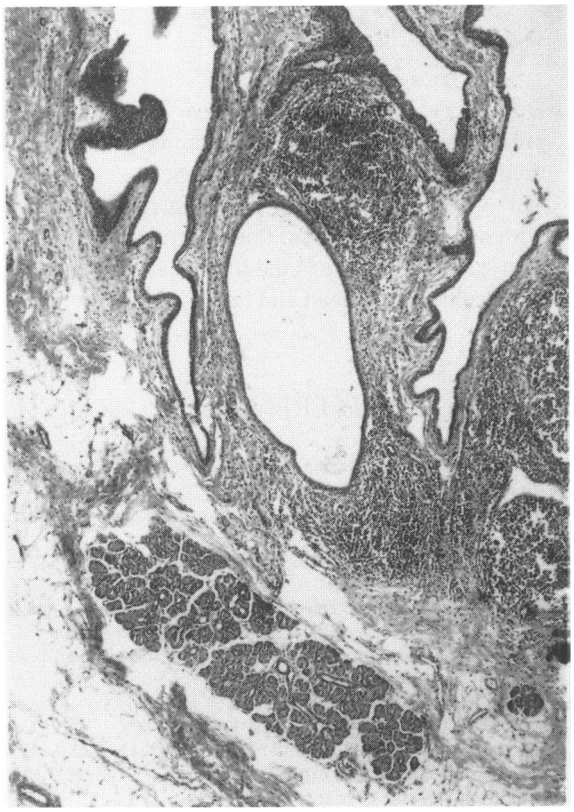

Figure 4. Low power view of bronchiolar gland arrangement reminiscent of endodermally derived tissue (hematoxylin and eosin stain).

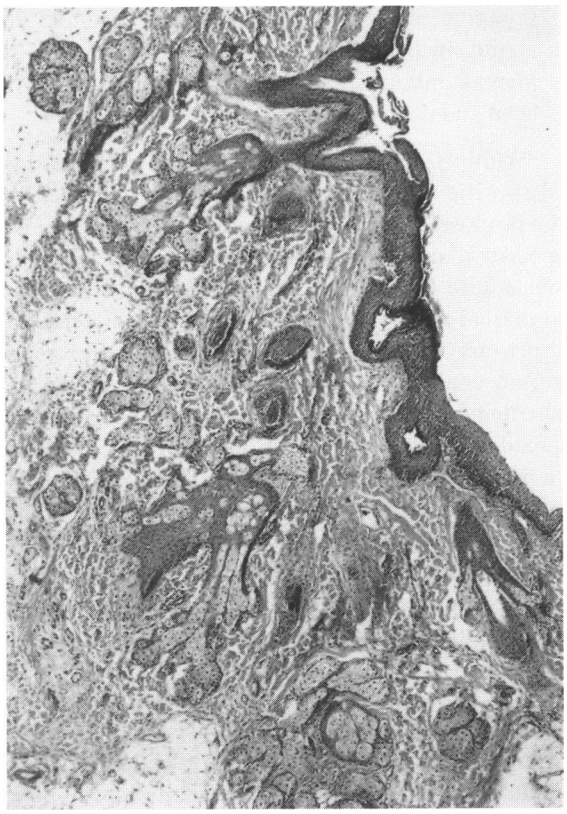

Figure 5. Seromucinous glands and metaplastic transitional epithelium are evident in this medium power view (hematoxylin and eosin stain).

ture fusion points in the calvarium, petrosquamous suture, nose, and orbit.

The epidermoid formation, recently described by Michaels[7] and Levenson et al,[8] is an interesting structure found imbedded within the tympanic membrane and middle ear anlagen from which congenital cholesteatomas are purported to arise. The epidermoid formation is found between 37 and 45 days gestation on the posterior aspect of the tubotympanic recess. Initially, it is in contact with the endodermal lining of the middle ear space. In later weeks, it enlarges and becomes active as the proposed organizer for the development of the middle ear cavity. It is always located in the anterosuperior quadrant near the bony tympanic ring. Usually disappearing by gestational week 33, the persistence of this structure could result in the classic findings of a pale anterior mesotympanic mass. While deemed responsible for epidermoid lesions in this area, the presence of this morphogenetically active site near structures of trigerminal origin could be the mechanism for the formation of dermoids and teratomas in this area also.

In reference to nomenclature, Batsakis[9] has defined a dermoid cyst as "an epithelial lined cavity with variable numbers of skin appendages (hair, follicles, sebaceous glands, sudoriferous glands, etc.)." Earlier pathology sources had more generally categorized these as being of ectodermal and mesodermal origin. These descriptions distinguish dermoids from teratomas (of trigerminal origin) and the teratoid cyst (trigerminal, but less differentiated and very rare in the head and neck).

In a review of 1495 dermoid cysts collected over 25 years at the Mayo Clinic, 6.94% were located in the head and neck.[1] This group was composed of orbital lesions in 49.5%, nasal lesions in 12.6%, submental or sublingual in 23.3%, and variously placed in the occipital, frontal, cervical, soft palatal, and lip regions in 14.6%. The classification system included

- *Congenital dermoid of the teratoma type,* specifically those arising from germinal epithelium. This grouping is most easily applied to those dermoids arising in the ovaries and testes. This also encompasses those lesions close to the site of either embryological neuropore (sacral or occipital).
- *Acquired implantation type,* or those inclusion dermoid cysts thought to be secondary to trauma. This group would include the pilonidal and distal extremity dermoids.
- *Congenital inclusion type,* or those arising along embryonic fusion sites such as the dorsal and ventral midline, calvarium, and branchial clefts. This

grouping would include the present case and those arising in the floor of mouth and sublingual, submental, intracranial, and epignathic lesions. Most head and neck dermoids fall into this group.

Within the category of congenital inclusion, several specific sites of origin in the head and neck have been proposed. Steel,[5] in his presentation of a hair bearing dermoid of the mastoid, suggests that the petrosquamous suture may be the site of inclusion of epidermal and dermal elements. Behnke[10] describes a dermoid of the petrous apex, citing Gacek's view that the foramen lacerum could be a source of embryonic mesodermal and ectodermal remnant tissue. Pensler[11] described the probable role of the fonticulus nasofrontalis functioning as a portal through which dural and dermoepidermal apposition and adherence might occur in the genesis of nasofrontal dermoids. Recently, Currarino and Rutledge[12] described temporoparietal dermoid cysts in two patients with subgaleal and epidural components. Dermoid cysts in the region of the anterior fontanelle have been described as well.

The lesion presented was noted to house a small amount of seromucinous tissue consistent with salivary gland. This most likely represents a choristoma of the middle and is not related to the dermoid component. The presence of enclaved salivary tissue in the middle ear is most likely another manifestation of the congenital inclusion phenomenon. Mischke et al,[13] in their description of the eighth case of middle ear choristoma, notes the association of this entity with aberrant seventh nerve anatomy and dehiscence with a high incidence of nerve transection during excision. In their review of the literature, all previous cases of middle ear choristoma were 9 years old or older.[13]

SUMMARY

1. Dermoid cysts of the middle ear are rare and can present behind an intact tympanic membrane, identical to congenital cholesteatoma.
2. This case represents the youngest patient reported with congenital dermoid (and choristoma) to date.
3. Congenital inclusion is the likely cause of most dermoid cysts in the head and neck.

REFERENCES

1. New GB, Erich JB. Dermoid cysts of the head and neck. Surg Gynecol Obstet 1937;65:48–55
2. Silberman R, Mendelson IR. Teratoma of the neck. Arch Dis Child 1960;35:159
3. Hinton J. Transactions of the Pathological Society of London, 1866;17:275
4. Fried MP, Vernick DM. Dermoid cyst of the middle ear and mastoid. Otolaryngol Head Neck Surg 1986;92:594–596
5. Steel A. Secretory otitis media due to a hair bearing dermoid of the mastoid cavity. J Laryngol Otol 1976;90:979–989
6. Howie TD. A case of dermoid or developmental cyst of the middle ear cavity. J Laryngol Otol 1962;76:62–66
7. Michaels L. Origin of congenital cholesteatoma from a normally occurring epidermoid rest in the developing middle ear. Int J Pediatr Otorhinolaryngol 1988;15:51–65
8. Levenson MJ, et al. Congenital cholesteatoma in children: An embryologic correlation. Laryngoscope 1988;98:949–955
9. Batsakis JG. Tumors of the Head and Neck, 2nd ed. Baltimore, Williams and Wilkins, 1979:227–228
10. Behnke EB. Dermoid of the petrous apex. Laryngoscope 1984; 94:779–783
11. Pensler JM. Craniofacial dermoids. Plastic Reconstr Surg 1988;82:953–958
12. Currarino G, Rutledge JC. Temporoparietal dermoid cysts with intracranial extension. AJNR 1988;9:385–387
13. Mischke RE, et al. Salivary gland choristoma of the middle ear. Arch Otol Head Neck Surg 1977;103:432–434